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Case Report

Malignant pelvic paraganglioma: A case report[☆]

Sara Azzabi Zouraq, M.D.^{a,b,c,*}, Meryem Haloua, M.D.^{a,b,c}, Badr Alami, M.D.^{a,b,c},
 Moulay Youssef Alaoui Lamrani, M.D.^{a,b,c}, Mustapha Maaroufi, M.D.^{a,b,c},
 Meryem Boubbou, M.D.^{a,b,c}

^aService de Radiologie, CHU HASSAN II de Fès, Fez, Morocco^bFaculté de médecine et de pharmacie de Fès, USMBA, Fez, Morocco^cLaboratoire de Neurosciences, Faculté de médecine et de pharmacie de Fès, USMBA, Fez, Morocco

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ABSTRACT

Pheochromocytomas arising from outside the adrenal glands are called paragangliomas and constitute a rare disease and can occur in the pelvic retroperitoneum. Symptoms of excess catecholamine production, as well as elevated urine vanillylmandelic acid levels and serum and urine norepinephrine levels, are highly diagnostic for paraganglioma. Imaging can be helpful for the diagnostic of these pelvic tumors, differentiating them from other pelvic masses. We hereby present a case of pelvic malignant paraganglioma.

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Introduction

The term paraganglioma refers to tumors arising from extra-adrenal chromaffin tissue (paraganglion cells) and can occur in the retroperitoneum (including the organ of Zuckerkandl), the head and neck region, and the urinary bladder. This entity is most seen in men between 20 and 40 years of age and is functional with associated hypertension in over three-fourths of cases [1]. Nonfunctional tumors may manifest with abdominal pain or a mass [1]. Unlike adrenal pheochromocytomas, approximately 30%–40% of extra-adrenal pheochromocytomas are malignant [2].

Paragangliomas can occur in isolation or as a part of multi-systemic disorders such as multiple endocrine neoplasia type

2a, neurofibromatosis, von Hippel–Lindau disease, or the triad of Carney (gastric leiomyosarcoma, pulmonary chondroma, and extra-adrenal paraganglioma) [3].

We hereby report a case of a pelvic malignant paraganglioma.

Case report

A 37 years old male patient was first admitted for signs of malignant hypertension.

The clinical examination found a PA at 180/80 at the time of admission. The patient also reported chronic pulsatile

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* Corresponding author.

E-mail address: sara.azzabizouraq@usmba.ac.ma (S.A. Zouraq).

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Fig. 1 – Two pelvis masses vividly and heterogeneously enhanced after contrast, with intimate relation with the iliac vessels.

headaches, palpitations and profuse sweating, what we call the “triad of MENARD”.

Laboratory data showed hyperlipidemia and high levels of serum neuron specific enolase (ng/mL) with a value of 5.900 ng/mL (normal value < 10), of serum catecholamines and urinary catecholamines including vanillylmandelic acid with a value of 23.800 mg/d (normal value 1.9-5.8) and homovanillic acid with a value of 13,600 mg/d (1.0-7.0).

A computed tomography (CT) was then performed, showing two pelvic masses in the internal iliac spaces, measuring 70 × 50 mm on the right and 50 × 40 mm on the left. These masses were heterogenous and spontaneously dense, vividly enhancing after contrast. Both masses showed intimate contact with the iliac vessels. The masse on the right showed invasion of the psoas muscle as well as the gluteus medius muscle. Both masses didn't show contact with the bladder. (Fig. 1)

The CT also showed an external iliac adenopathy measuring 40 × 25 mm.

A biopsy of the adenopathy was performed which was compatible with a neuroendocrine tumor.

Additional immuno-histochemistry was then performed, confirming the diagnosis of malignant paraganglioma.

Further investigations were necessary to look for other localizations, especially looking for Multiple neuroendocrine neoplasia, which all came back negative.

The two masses were judged non resectable surgically due to their intimate contact with the iliac vessels.

The patient underwent radiotherapy, with a dose of 60 Grey (Gy) (2Gy in 30 sessions).

At the 6 months follow-up, the patient showed an improvement of his hypertension, with a PA of 140/80.

The laboratory control showed the persistence of high level of urinary catecholamines.

The control CT showed a regression of the two masses previously described (Fig. 2).

The patient is currently still undergoing radiotherapy treatment.

Discussion

Generally, pheochromocytomas arising from outside the adrenal glands are called paragangliomas and constitute a rare disease. Approximately 18% of pheochromocytomas are paragangliomas [4,5]. Malignant paragangliomas are seen in 29%-40% of cases, which is higher than that of adrenal pheochromocytomas (2%-11%) [4,5]. Functional paragangliomas are frequently malignant and associated with a high incidence of persistent or recurrent disease.

Tumor size of over 5 cm and occurrence of invasion or metastatic disease are strong predictors of malignancy [6]. Approximately 10% of patients with retroperitoneal paraganglioma that present with back pain or a palpable mass have a distant metastasis at diagnosis. The metastatic potential of retroperitoneal paraganglioma is higher and ranges from 20% to 42% [6].

Symptoms of excess catecholamine production, as well as elevated urine vanillylmandelic acid levels and serum and



Fig. 2 – Regression of the two iliac masses after 6 months of radiotherapy.

urine norepinephrine levels, are highly diagnostic for paraganglioma [7]. Percutaneous biopsy of a hypervascular pelvic retroperitoneal mass without adequate laboratory evaluation and prebiopsy preparation (such as pharmacologic blockade) should be avoided, since it can result in hypertensive crisis or even death.

In CT, paraganglioma appears as a well-defined hypervascular mass in the pelvic retroperitoneum along the course of the common iliac vessels. These tumors may be heterogeneous with foci of calcification. Evidence of local invasion and metastasis may be seen in malignant tumors.

At MR imaging, pheochromocytoma appears as areas of low T1 and high T2 signal intensity and shows avid enhancement following the administration of gadolinium-based contrast material [3]. The sensitivity of CT and MR imaging for the detection of extraadrenal pheochromocytoma is approximately 90% [3]. MIBG scintigraphy has traditionally been used to localize adrenal pheochromocytoma as well as extraadrenal paraganglioma.

The differential diagnosis for paraganglioma in the pelvic retroperitoneum includes

Nerve sheath tumors:

- Schwannoma: manifest as well-defined hypoechoic masses at US and as hypoattenuating masses at CT. Areas of cystic degeneration, calcification, or hemorrhage may also be seen at CT. At MR imaging, schwannomas appear as well-defined masses with heterogeneous low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. A target-like pattern consisting of peripheral high signal intensity (representing Antoni B areas) and central low signal intensity (representing Antoni A areas) may also be seen on T2-weighted images [8].
- Neurofibromas: manifest as well-defined, homogeneously hypoechoic masses at US. Plexiform neurofibromas are hypoattenuating relative to soft tissue at CT due to the presence of lipid-rich Schwann cells, adipocytes, and myxoid change [9]. Pelvic plexiform neurofibromas tend to be large and may appear as hypoattenuating, hypo-vascular

infiltrative masses or as intensely enhancing soft-tissue masses [10].

Hyper vascular lymphadenopathy (metastases or Castleman disease), and other hyper vascular soft-tissue sarcomas.

Primary retroperitoneal tumors of mesodermal origin, specifically lipomatous tumors, may be differentiated from paragangliomas on CT only if fat can be identified within the tumor.

Poorly differentiated liposarcomas with little or no fat, regardless of whether they are solid or necrotic [11–13] are difficult to distinguish from paragangliomas. This is also true of leiomyosarcomas. However, a necrotic or cystic liver metastasis would be more likely to occur in leiomyosarcomas than in paragangliomas [14]. Malignant fibrous histiocytomas arising near the kidney or adrenal gland would be indistinguishable from renal hilar paragangliomas [15].

Patient consent

All the authors certify that the patient has consented to the publication of the case.

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